CORONARY ARTERIES AND RIGHT VENTRICULAR HISTOLOGY IN HYPOPLASTIC LEFT HEART SYNDROME

Jeanne M Batta, MD, Sheng-Liang Chen, MD, Maria E Guttenberg, MD, William F Norwood, MD, PhD, F.A.C.C., Paul M Weinberg, MD, F.A.C.C.
The Children's Hospital of Philadelphia, Philadelphia, PA.

It has been suggested that in the subgroup of patients with hypoplastic left heart syndrome(HLHS) with a patent mitral valve and an atretic aortic valve, the coronary arteries are sufficiently abnormal to warrant referral for heart transplantation in lieu of reconstructive surgery. We evaluated 150 pathologic specimens with HLHS with aortic valve atresia or hypoplasia, 95 with a patent but hypoplastic mitral valve (MH), 54 with mitral atresia (MA) and 1 with malaligned atrioventricular canal, to determine if RV preservation is significantly compromised in any of the anatomic subgroups of HLHS. The age at death ranged from 1 day to 28 months, median age 8 days. Cause of death included preoperative hemodynamic (76), intraoperative or perioperative (47), late hemodynamic (>30 days postoperatively) (3), sudden death/arrhythmia (1), respiratory , infection or other (15). There was no significant difference among the anatomic subtypes as to age at death or cause of death. In 74 pts, 87 surgical procedures had been performed including Stage I Norwood procedure (66), modified Fontan procedure (11) and other (10).

In all specimens, the coronary arteries were examined macroscopically. Gross coronary anomalies included arteriocameral fistulae (6), single right coronary artery (4), single left coronary artery (1), and tortuosity (1). Only the presence of arteriocameral fistulae was different among the subgroups, associated significantly with MH. Histologic sections were obtained from 26 hearts, 11 with MH, 10 with MA, and 5 control hearts, including sections of RV, LV and coronary arteries with adjacent myocardium. . Measurement of histologic sections showed no significant increase in the ratio of coronary arterial wall thickness to lumen diameter in either the MH or MA groups. While ischemia and endocardial fibroelastosis were noted in the LV of the MH group and not in the MA group, the RV histology was similar in both, and it appeared unaffected by advancing age.

We conclude that in the absence of significant differences in RV myocardium between MH and MA groups, there is no subgroup of HLHS which is unfavorable for reconstructive surgery.

THE EFFECTS OF PENTOXIFYLLINE (TRENTAL®) ON BLOOD FLOW, VISCOSITY, AND OXYGEN TRANSPORT IN YOUNG ADULTS WITH INOPERABLE, CYANOTIC CONGENITAL HEART DISEASE.

William Berman Jr., M.D., F.A.C.C., Norman B. Berman, M.D., Stephen C. Wood, PhD, and Dorothy Patnak, PhD, University of New Mexico Albuquerque, NM

Four polycythemic young adults (13-28 years old) with inoperable, cyanotic congenital heart disease (CHD) due to congenitally abnormal and hypoplastic pulmonary arteries (1) or pulmonary vascular disease(3) were treated for 12 weeks with 20 mg/kg/day of pentoxifylline in an attempt to increase pulmonary blood flow by improving red blood cell deformability and reducing whole blood viscosity. Measurements made for 6 weeks before, 12 weeks during, and 6 weeks after treatment showed: a fall in O_2 consumption from 185 to 156 ml/min/M^2; a rise in arterial (75% to 83%) and mixed venous (42% to 51%) O_2 saturation; a rise in the effective pulmonary blood flow index from 1.4 to 1.7 L/min/M^2; and a rise in systemic O_2 transport (405 to 469 ml/min/M^2), with an attendant fall in the O_2 extraction coefficient from 47% to 37%. Systemic blood flow, hematocrit, & right to left intracardiac shunting did not change. Pentoxifylline decreased whole blood, but not plasma, viscosity at all hematocrits over a range of shear rates. Pentoxifylline also shifted the oxy-hemoglobin dissociation curve to the right, increasing the already elevated P_{50} from 34 to 36 torr. Two patients had significant nose bleeds during treatment, one requiring hospitalization for nasal packing. Pentoxifylline may improve oxygenation in some patients with inoperable CHD, but the risk of bleeding must be considered.

EARLY AND LATE RESULTS OF THE FONTAN PROCEDURE FOR DOUBLE-INLET LEFT VENTRICLE: THE MAYO CLINIC EXPERIENCE.

Douglas D. Mair, M.D., F.A.C.C., Donald J. Hagler, M.D., F.A.C.C., Paul R. Julsrud, M.D., Francisco J. Puga, M.D. F.A.C.C., Hartzell V. Schaff, M.D., F.A.C.C., Gordon K. Danielson, M.D., F.A.C.C., Mayo Clinic and Mayo Medical School, Rochester, MN

Between 1977 and March 1989, 155 pts with double inlet left ventricle (DILV) had the Fontan procedure performed at the Mayo Clinic. Age at operation ranged from 1 year to 41 years (median = 10 years). Operative mortality from 1977 through 1980 (39 pts) was 21% but for 1981 through 1989 (116 pts) was 9%. There have been 17 late deaths secondary to reoperation (8), progressive myocardial failure (5), sudden dysrhythmia (3), and bleeding varices (1). Follow-up of 6 months to 11 years (mean = 4.9 years) in 111 pts revealed 88% to be in good or excellent condition and 12% to be fair or poor.

Operative and late mortality were directly related to preoperative mean pulmonary artery pressure (PAP) with the combined overall mortality being 12% in those with a mean PAP of 20 mm Hg or less and 41% in those with mean PAP greater than 20 mm Hg. Combined operative and late mortality was not related to age at operation.

The Fontan operation can presently be performed with an operative risk of less than 10% in properly selected pts with DILV and late results are encouraging when contrasted with the clinical course of these pts before this operative approach was utilized.

EXERCISE PERFORMANCE WITH RATE RESPONSIVE PACING IN COMPLEX CONGENITAL HEART DEFECTS.

Stephen M. Paridon MD, Peter P. Karpawich MD, William W. Pinsky MD FACC. Division of Cardiology, Children's Hospital of Michigan, Wayne State University, Detroit, MI.

Benefits of ventricular rate responsive (VVI-R) pacing have been well described in the adult. Its use at the atrial level (AAI-R) or in congenital heart defects has not been reported previously. We evaluated the effects of both R pacing modes vs demand mode (rate 70/min) on exercise performance in 11 postoperative pts with congenital heart defects: AAI-R in 3 pts (Mustard 2, ASD) and VVI-R in 8 pts (truncus arteriosus 1, Fontan 2, Mustard 1, VSD 1, Ebstein's 1, congenital heart block 1, ASD 1). Activity settings: Low rate 70/min, maximum R-rate of 150/min. Each pt was exercised to maximum volition using a treadmill protocol with pacing mode randomized to demand mode, nominal activity sensitivity and then maximum activity sensitivity. Heart rate (HR), oxygen consumption (VO_2), work rate (WR), and respiratory exchange ratio (RER) were monitored continuously. Results are summarized below:

Pacing Mode	Max HR (BPM)	Max VO_2 (ml/kg/min)	Max WR (Watts)	(% Predicted)	Max RER
Demand	109	29	161	(90)	1.24
Nom. Activity	130	29	158	(92)	1.31
Max. Activity	141*	36#	170	(106)	1.17

Different from Demand *p=0.01 # p=0.05

We conclude that in pts with congenital heart defects 1) exercise capacity as measured by work rate is generally good in all pacing modes; 2) However, rate responsive pacing results in a significant improvement in maximum heart rate and maximum VO_2; 3) Maximum rather than nominal activity settings may be required to achieve improved chronotrophic response with activity pacing in these patients.